Case Report

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Idiopathic bilateral occlusion of Foramen of Monro treated by septostomy with unilateral foraminoplasty: a rare case report

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Introduction: Idiopathic occlusion of the Foramen of Monro is extremely rare in adults. The occlusion is classified into four types, with the first being the most infrequent. This condition induces noncommunicating hydrocephalus with the ensuing increased intracranial pressure symptoms. Headache is usually the chief complaint.

Presentation of the case: The authors present a case of a 28-year-old female who presented with a chronic headache that was unresponsive to analgesics. No other neurological deficits were present. Fundoscopic examination revealed the presence of bilateral papillary edema. Computed tomography scan results showed bilateral enlargement of the lateral ventricles of the brain. A subsequent MRI scan ruled out secondary causes of occlusion, such as colloid cysts, meningiomas, or choroid plexus tumors, which entailed an idiopathic etiology. Treatment options include ventriculoperitoneal shunt insertion and septostomy with foraminoplasty. The former option is currently the treatment of choice, yet it is notorious for its ramifications, including foreign body reaction, breakage, and mechanical problems. The latter option is free of these risks; however, it requires meticulousness and precision to avoid damaging the fornix, which leads to impaired memory function.

Conclusion: Septostomy with unilateral foraminoplasty could yield better outcomes if it is performed fastidiously.

Keywords: Monro Foramen Occlusion, Hydrocephalus, Septostomy, Foraminoplasty, Ventriculoperitoneal shunt, Case Report

Introduction

Foramen of Monro (FM), which was first described *by Alexander Monro Secundus (1733–1817)*, is a conduit which bridges the lateral ventricle of the brain with the third ventricle on either hemisphere[1].

The etiology of FM occlusion includes numerous disorders including colloid cysts, meningiomas, subependymomas, hypothalamic gliomas, central neurocytomas, choroid plexus tumors, inflammatory conditions (brain abscesses and ventriculitis), vascular malformations, basilar artery aneurysm, infections (particularly TORCH infections which can induce inflammation and scarring), and iatrogenic complications[2–4].

In extremely rare cases, no explicit etiology can be identified, which entails an idiopathic causality. Only 26 cases have been documented in the literature[2,3]. Even more scarcely, the occlusion can affect both lateral ventricles of the brain, making the case exceedingly rare, with only 17 cases described[2,3]. Therefore, an idiopathic FM occlusion is an extremely rare phenomenon[2].

Bilateral FM occlusion leads to noncommunicating (obstructive) hydrocephalus[1].

Clinically, symptoms of increased intracranial pressure (ICP) are present, with headache being the most prominent manifestation[5].

Regarding treatment, several surgical procedures are to be considered, including septostomy, foraminoplasty, and the placement of ventriculoperitoneal (VP) shunt[2,6]. In case of unequivocal anatomical landmarks, septostomy and/or foraminoplasty are the treatment of choice. Conversely, if anatomical distortion is confirmed, a VP shunt should be considered[7].

This paper presents a case of idiopathic bilateral FM occlusion, which was treated with septostomy with unilateral foraminoplasty.

HIGHLIGHTS

- Idiopathic bilateral occlusion of Foramen of Monro is extremely rare in adults, with less than 30 cases reported in the literature.
- Septostomy and unilateral foraminoplasty are reported successfully once in the literature, and to the best of our knowledge, this is the second case to be reported.
- The current first-line treatment is ventriculoperitoneal insertion; however, if anatomical landmarks are recognized clearly, septostomy with unilateral foraminoplasty could be a better choice.

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This case has been reported in line with the SCARE (Surgical Case Report) criteria[8].

Presentation of the case

A 29-year-old female presented to the emergency department of Al-Mouwasat University Hospital in June of 2023, complaining of a chronic bilateral headache that had persisted for the past 2 years. Initially, the headache manifested once a week and responded well to analgesics. The headache-free intervals gradually shortened until it reached the point where the headache would awaken the patient from her slumber and last for several consecutive hours, or occasionally all day long. Ultimately, the use of analgesics including non-steroidal anti-inflammatory drugs, yielded no effect.

Concurrently, the patient experienced typical symptoms of increased ICP including nausea, vomiting, and blurred vision.

The patient's medical history involved recurrent miscarriages; however, it is void of head trauma or infection and any form of substance intake including alcohol and tobacco.

Both physical and neurological examinations yielded normal results and laboratory tests were within normal limits as well. Bilateral papilledema was detected via fundoscopy. A subsequent contrast-enhanced computed tomography (CT) scan indicated biventricular enlargement consistent with hydrocephalus. A prophylactic Zolamid dose was administered with the objective of mitigating the increased ICP. An MRI was performed to further investigate the prior finding, which confirmed the bilateral hydrocephalus and ruled out any mass-occupying lesions as well. (Fig. 1A, B).

Thus, a strong suspicion of an idiopathic bilateral occlusion of the FM was established. Endoscopic surgery was chosen. A right-sided Kocher's point, 3 cm lateral to the midline, was located and a typical frontal burr hole was made. A severe occlusion of FM was noted confirming the initial diagnosis (Fig. 2A, B).

Consequently, a septostomy and foraminoplasy were performed and the resultant restoration of cerebrospinal fluid (CSF) was observed (Fig. 2C).

Following the procedure, rapid resolution of symptoms was reported. The patient was monitored for 3 days in the hospital, during which she received prophylactic antibiotics (ceftriaxone and vancomycin) and a 24-hour postoperative MRI, which revealed a reduction in the size of the lateral ventricles and normalization of the CSF circulation (Fig. 1C).

The patient was discharged with no reported complications.

Discussion

FM occlusion is ascribed to numerous disorders, mainly tumors, including colloid cysts, which are the most common masses that affect the FM with an incidence rate of 0.2–2% of all intracranial tumors, infections, hemorrhages, or aplasia[1,2].

Failure to discern an explicit etiology of the occlusion leads to classifying the condition as idiopathic, which is a rare occurrence.

Idiopathic occlusion of Foramen of Monro (IOFM) is categorized into four types; types 1 and 2 are bilateral whilst types 3 and 4 are unilateral; thus, they are associated with septum deviation and a resultant contralateral functional occlusion of FM.

Additionally, types 1 and 3 describe true stenosis of FM, whereas types 2 and 4 involve membranous occlusion of FM[2].

In the majority of cases including ours, the type of the occlusion can be determined intraoperatively. However, Mizrahi *et al.* put forward the utilization of high-resolution MRI with multiplanar constructions at the level of FM for preoperative identification of the type[2,9].

This paper features a bilateral true stenosis of FM (type 1).

The condition seems to affect young adults with a mean age of 33 years old; however, our patient was 29[9].

Patients suffering from IOFM exhibit symptoms associated with elevated ICP including headache, vomiting, gait disturbance, blurred vision, and memory impairment[2].

Similarly, our patient presented with a chronic headache and blurry vision.

Other symptoms have been reported in the literature as well, De Bonis *et al.* mentioned urinary incontinence, deceleration of

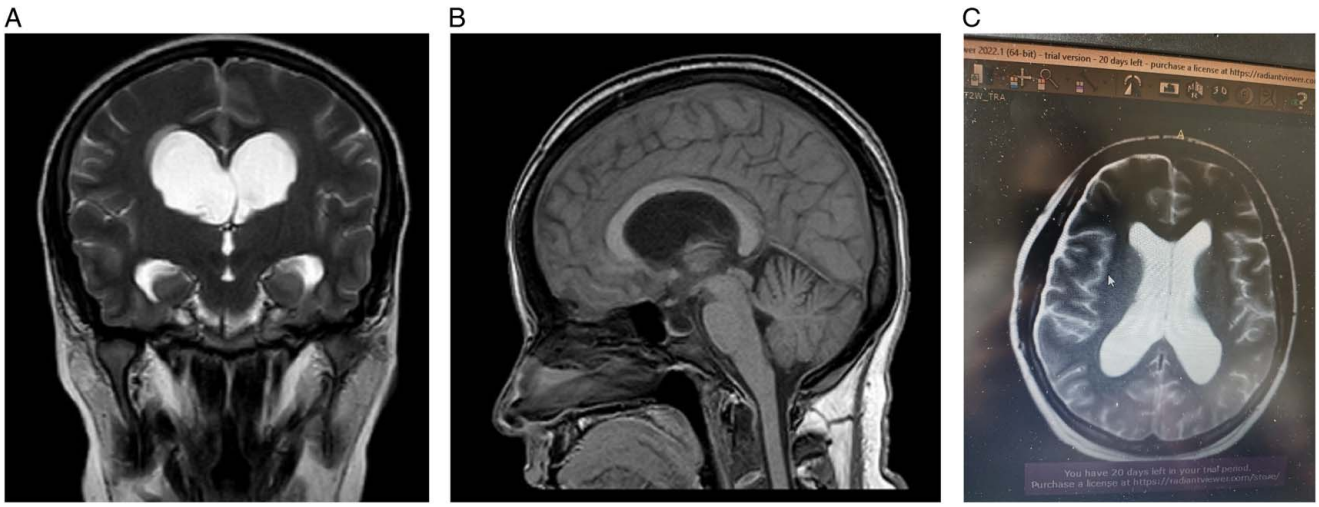

Figure 1. Nonenhanced MRI findings. (A, B) Coronal and sagittal planes demonstrate severe isolated dilatation of the lateral ventricles on T2-weighted and T1-weighted images, respectively, with normal appearing third and fourth ventricles. (C) The axial plane highlights the reduction in the severity of the aforementioned dilatation after treatment with spetostomy and unilateral foraminoplasty.

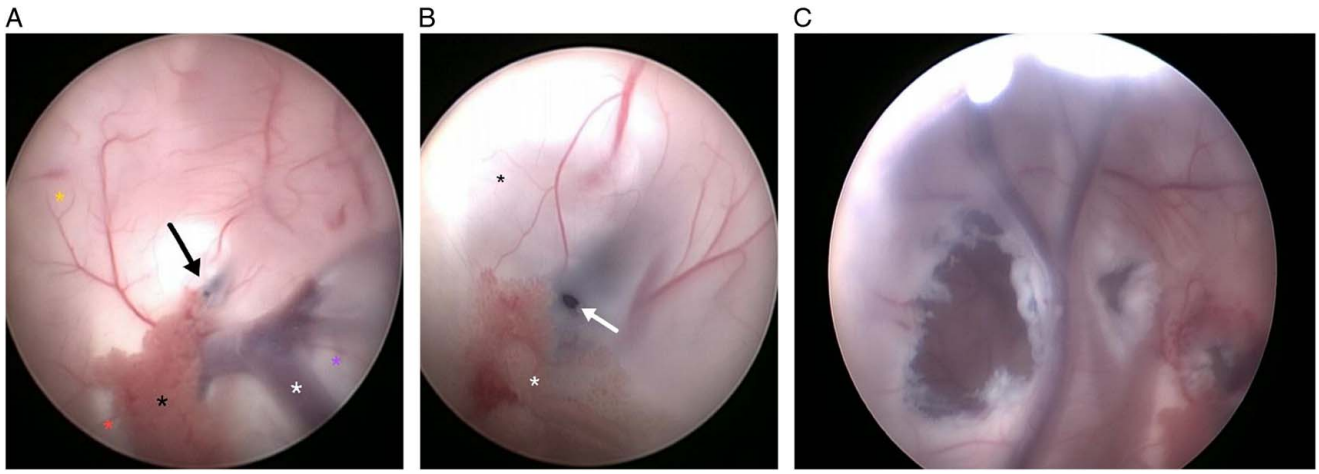

Figure 2. Intraoperative pictures illustrating the anatomical landmarks of the area and the stenotic foramen. (A) The true stenosis of the Foramen of Monro can be visualized clearly (black arrow). The choroid plexus is seen as well (black asterisk) next to the fornix (red asterisk) and the thalamostriatal vein (white asterisk). Adjacent to the vein, the thalamus can be seen (blue asterisk). Lastly, the septum pellucidum can be observed (yellow asterisk). (B) A better view of the stenotic foramen of Monro (white arrow). The choroid plexus can be visualized as well (white asterisk) along with the septum pellucidum (black asterisk). (C) Postoperative picture showing the septostomy and the foraminoplasty of the Foramen of Monro.

thought, and asthenia, Freudenstein *et al*. reported vertigo as one of the symptoms, and Maldonado *et al*. described visual flashes and memory loss. However, our patient reported none of these symptoms[3,10–12].

In regards to signs, papillary edema is detected in some cases; nevertheless, it is not always present. In our case, the patient was found to have bilateral papillary edema upon fundoscopy.

In terms of diagnosis, identification of bilateral ventriculomegaly with a shrunken third ventricle on CT scans can imply the diagnosis of IOFM; nevertheless, a consecutive MRI utilizing T1-weighted and T2-weighted studies, FLAIR (fluid-attenuated inversion recovery), and DWI (diffusion-weighted imaging) protocols is necessary to rule out secondary causes of FM occlusion[2,9].

Regarding treatment, two options are to be considered when managing IOFM: conservative and surgical approaches.

The former approach could be a viable option in the absence of neurological deficits or signs of increased ICP, especially papillary edema. In such cases, analgesics and acetazolamide could be prescribed with close monitoring for the development of any ICP-related symptoms. Mizrahi *et al*. reported two cases successfully treated conservatively; one of them was a unilateral occlusion of FM whilst the other was bilateral. They were followed up for 3 years and 10 months, respectively, and the patients maintained an asymptomatic course[2,9].

On the contrary, our patient failed to respond to analgesics, which necessitated the surgical approach. Failure to respond to the conservative approach is one indication for considering surgery, and the other three indications are severe acute or subacute clinical presentation, the presence of neurological deficit that is ascribed to elevated ICP, and signs of ICP such as papillary edema[2].

The type of surgical approach is reliant on the IOFM. Type 1 IOFM has been reported to be optimally managed with VP insertion and septostomy; however, endoscopic procedure has also been reported to yield excellent outcomes. The endoscopic procedure involves a unilateral foraminoplasty and a septostomy[2,9].

On the one hand, endoscopic treatment has several advantages over VP insertion including direct visualization of the underlying cause, minimal invasion, a low-risk profile, and the possibility of obtaining a biopsy specimen if needed. On the other hand, the endoscopic procedure has two main risks; interventricular hemorrhage, and damage to the fornix, which leads to memory impairment. However, these complications are deemed rare[2]. Therefore, our patient underwent unilateral foraminoplasty with septostomy.

Migliorati *et al*. reported a case where a patient had to undergo foraminoplasty with septostomy due to VP insertion inadequacy and its ensuing complications. The patient immediately developed short-term memory impairment due to fornix damage, which was the result of poor visualization of anatomical landmarks[6]. Thus, certain anatomical landmarks should be identified before performing septostomy including FM, the choroid plexus, the thalamostriatal vein, and the anterior caudate vein. In case of an inability to recognize these landmarks, the procedure should be aborted[7].

It is important to mention that type 1 IOFM has only been reported six times in the literature; only one of them was treated successfully with septostomy and unilateral foraminoplasty without the need for a VP shunt[2].

Thus, and to the best of our knowledge, this paper is the second in the literature that reports a successful management of type 1 IOFM with septostomy and unilateral foraminoplasty. Interestingly, some patients require the placement of a shunt due to failure of the endoscopic procedure[2].

Types 2, 3, and 4 could all be optimally approached endoscopically with excellent results[2].

Conclusion

IOFM is a rare occurrence that has been classified into four types. Type 1 is thought to be best managed with VP insertion; however, with a precise understanding of the anatomical landmarks of the ventricles and meticulousness during the procedure, septostomy

with foraminoplasty could yield better results, especially when considering the low-risk profile and the lessened complications in comparison to VP insertion.

Ethical approval

Not applicable.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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None.

Author contribution

G.I.A.S.: wrote all paragraphs of the research article; A.M.W.: extracted the references and drafted the manuscript; A.A.: collected the data and participated in academic writing; A.M.A. and R.N.Z.A.: are the surgeons who made the diagnosis and the surgical intervention and revised and improved the research paper. All authors read and approved the final version of the manuscript.

Conflicts of interest disclosure

There are no conflicts of interest.

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Data availability statement

Since all data are included in the manuscript, there is no need for data sharing.

Provenance and peer review

Not invited.

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